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Giant paraganglioma of the parapharingeal space in a woman

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Abstract: The case report of a 57 years patient with history of hypertension is described who was assisted in the Head and Neck Surgery Service of the National Institute of Oncology and Radiobiology in Havana, referred from the Otolaryngology Service of her head hospital due to clinical manifestations of dysphagia and suspected clinical diagnosis of a carotid paraganglioma. After the physical exam and the pertinent complementary studies, the case was discussed by the members of a multidisciplinary team and the existence of a paraganglioma in the parapharingeal space was confirmed, therefore it was decided to carry out surgical treatment. The clinical course was satisfactory, and 2 years after the surgical procedure there were no sequels.

Key words: paraglanglioma; carotid body tumor; parapharingeal space

1 Introduction

Paragangliomas are rare lesions in the head and neck that arise from a tissue called paraganglion. They are of neuroectodermal, adrenal, or extra-adrenal embryological origin, slow-growing, and benign in nature, although some have characteristics similar to those of malignant tumors [1].

In 1743, Von Haller [2] detailed the carotid body, and in 1772, Neubauer produced the first diagram of this structure. In 1891, Marchand reported the first paraganglioma of the carotid body and described it as a flattened structure measuring 2.5 by 5 mm, located on the inner surface of the upper end of the common carotid artery, near its bifurcation into the internal and external carotid arteries [3]. It is also claimed to be a chemoreceptor organ, supplied by the glomus or ascending pharyngeal arteries, whose innervation is provided by cranial nerves IX, X, and XII, which form the carotid sinus nerve or nerve of Hering [4].

Later, in 1971, Shamblin proposed a classification of carotid body paragangliomas and divided them into three grades [5,6]:

1. Small lesions, loosely attached to the carotid arteries (easily dissectable).
2. Larger lesions that partially surround the carotid arteries and are attached to the vascular adventitia (the most common).
3. Lesions that completely surround the carotid bifurcation (higher incidence of complications).

On another note, paragangliomas can form at any age, but they are most often diagnosed in adults between the ages of 30 and 50. In most cases, the cause is unknown, but in others it is related to a set of genetic mutations inherited from

parents to children [7].

Paraganglioma cells often secrete hormones called catecholamines, such as adrenaline, whose function is to trigger an alarm reaction. This can cause high blood pressure, rapid heart rate, sweating, headache, and tremors.

The treatment of choice is surgery, which poses a challenge for surgeons due to the neurovascular structures present. Given the rarity of parapharyngeal paragangliomas, it was decided to report the case to the international medical community.

2 Clinical case

We present the clinical case of a 57-year-old female patient with a history of hypertension, controlled with regular treatment, who attended the Otorhinolaryngology Service of her local hospital complaining of difficulty swallowing and a pulsatile, movable nodular lesion measuring 5 cm in diameter in the lateral region of the neck (levels II and III).

3 Physical examination

Examination of the aerodigestive tract revealed an increase in volume, with a tumor-like appearance, on the left side wall of the pharynx, which was hard and elastic in consistency, extending to the base of the tongue and partially occluding the lumen of the oropharyngeal cavity. Given the complexity of the case, the patient was referred to the Head and Neck Surgery Service of the National Institute of Oncology and Radiobiology in Havana with a clinical diagnosis of suspected carotid paraganglioma, where she underwent additional hematological, blood chemistry, and coagulation tests, the results of which were normal.

4 Other studies of interest

1. Simple and contrast-enhanced computed tomography of the head and neck (3 mm slices): A hyperdense, heterogeneous image, resembling a tumor, measuring 62 by 58 mm and with a density of 41 HU in a simple series, was observed occupying the left side of the pharynx and protruding into the lumen, with almost total stenosis of the lumen and extension to the base of the tongue (exceeding the midline), in contact with the ipsilateral prevertebral region and vascular structures, but with a fatty interface, as well as contrast uptake up to 169 HU and hypodense areas within it.
2. Diagnostic four-vessel CT angiography: The presence of a tumorous and hypervascular lesion was confirmed in the left cervical region, in front of the carotid artery, which displaced it posteriorly and received its main afferent through the ascending pharyngeal artery and the fine branches dependent on the external carotid artery on the same side.
3. Endovascular procedure: A guide catheter was placed in the ascending pharyngeal artery and navigated with a Sonic 1.2 microcatheter and a Hybrid 0.07 microguide to access it distally. PHIL-25% (1.2 cm^3) was injected, and the tumor afferents dependent on this branch were successfully obliterated, which constituted practically 100% of the lesion. It should be noted that the follow-up angiography revealed only a minimal late-phase venous blush. (Figure 1).

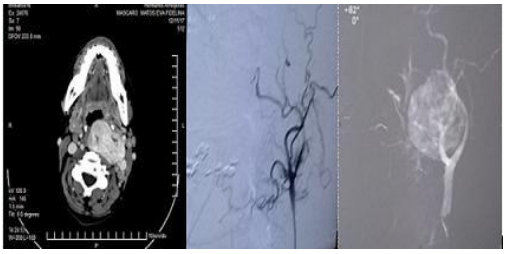

Figure 1. CT angiography and embolization

Fine-needle aspiration cytology was performed, which was negative for neoplastic cells, while the cytological smear showed epithelioid cells with eosinophilic cytoplasm compatible with paraganglioma.

The case was discussed by the members of a multidisciplinary team, who confirmed the existence of a type II carotid paraganglioma (Shamblin classification) and decided to proceed with surgical treatment.

5 Surgical procedure

Forty-eight hours after selective embolization of the tumor, the patient was transferred to the surgical unit. In the supine position with the neck in hyperextension, a modified Edgerton incision was made. The large vessels of the neck were identified and controlled, as were the nerve structures related to the lesion, such as the hypoglossal, vagus, and superior laryngeal nerves. Careful subadventitial dissection of the tumor was then performed in a caudocephalic direction, and meticulous hemostasis was achieved until excision was completed (Figure 2).

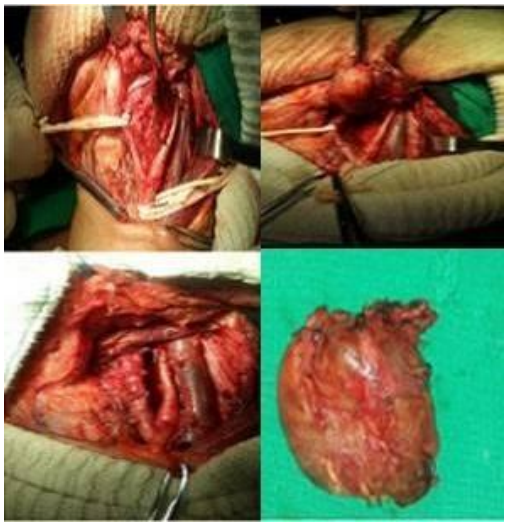

Figure 2. Surgical resection of the tumor and sample for histological study

The histological result of the excised portion confirmed the diagnosis of carotid body paraganglioma.

Two years after the surgical procedure, the patient continued to show excellent progress.

6 Comments

Paragangliomas are rare tumors, with a prevalence of 1 in every 30,000 to 100,000 individuals in the general population. The most common type is carotid body paraganglioma, which accounts for approximately 60-78% of cases. This is the only disease that affects the carotid body, whose presentation in the parapharyngeal space is very unlikely, as it represents between 0.5-0.8% of all head and neck tumors. Of these, the most common are those of the accessory salivary glands (40-50%), with neurogenic tumors (17-25%) and paragangliomas (10-15%) occurring less frequently [8,9].

The preferred treatment is excision, so it is useful to determine the vascular flow of the lesion by means of angiography or angiotomography. The choice of therapeutic modality depends mainly on the location and size of the lesion, the age and health status of the patient, as well as the preferences of the patient and surgeon.

Similarly, for paragangliomas larger than 2 cm, preoperative embolization is recommended to reduce the size and vascularization of the tumor, thereby improving visibility in the surgical field and reducing blood loss and the risk of injury to vascular and nerve structures. Once this procedure has been performed, excision is recommended within the first 48 hours to prevent revascularization.

In this regard, other authors [10] suggest radiotherapy as an alternative treatment to prevent tumor progression when surgery is contraindicated. On the other hand, the complication rate is high, with a mortality rate of 1-2% and a morbidity

rate of 40%, including cranial nerve injury, hemorrhage, brachiocephalic syndrome and cerebrovascular accident , among others. Furthermore, the involvement of nerve structures is directly proportional to the location during the surgical procedure, the size, and the need for vascular reconstruction.

In this clinical case, the authors consider the extension of the tumor to the parapharyngeal space to be relevant and that no complications have occurred in 2 years of follow-up.

Given the complexity of the surgical procedure for this type of injury, it is important to understand the therapeutic principles involved in order to ensure the patient's quality of life.

Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

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